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Editorial Comment

The Various Therapeutic Approaches to Aortic Coarctation: Is It Fair to Compare?*

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Aortic coarctation is defined as a reduction in the diameter of the aorta at the junction of the aortic isthmus and the descending thoracic aorta. However, this definition belies the deceptively complex physiologic and morphologic variability of this lesion. This observation is underscored by the fact that despite >50 years of surgical and medical management of coarctation, a complete understanding of its pathophysiologic characteristics and a consensus on the optimal form of therapy continue to elude us.

Background. The variability and complexity of this lesion can be appreciated when one considers several important facts: 1) Aortic coarctation can present at any time between the first few hours of life and early adulthood. 2) The presence or absence of a number of associated lesions has important implications. Associated lesions include patent ductus arteriosus; proximal tubular isthmus and aortic arch hypoplasia; associated intracardiac defects, ranging from ventricular septal defect to complex intracardiac anomalies resulting in single-ventricle physiology; descending aortic collateral vessels; and aneurysmal degeneration of the aorta. Attempts have been made to categorize these associated anomalies using the continuum from the "infantile" form to the "adult" form of aortic coarctation. There is a certain validity to this continuum in which the association of patent ductus arteriosus, tubular hypoplasia and intracardiac defects (infantile form) give way to a more discrete coarctation ring or hourglass deformity without patent ductus arteriosus or intracardiac defects but with secondary development of arterial collateral vessels to the lower body aorta and degenerative aortic changes (adult form).

This morphologic variability presents a number of very practical difficulties, among them the difficulty of defining or grading the severity of the coarctation. For example, in the infantile form of the lesion the presence of the patent ductus arteriosus alone not only prevents a meaningful physiologic assessment of the obstruction using the pressure gradient, but it also introduces the additional problem of metamorphosis: that is, the changing state of contraction of the ductus arteriosus itself is a major determinant of the degree of coarctation (the ductal sling concept). As a result, the severity of aortic coarctation is often defined by a somewhat subjective combination of physiologic variables (pressure gradient) and morphologic variables (length and diameter of the narrowing in the aorta). Even in more "adult" forms of coarctation, this problem exists. The presence of well developed collateral vessels often makes assessment of the severity of the coarctation difficult, in that the pressure gradient may become reduced as a result of the development of efficient collateral flow. The case of a completely occluded aortic lumen associated with little or no pressure gradient between the ascending and descending aorta is the extreme example of this phenomenon.

The variability of coarctation, in conjunction with other factors, has spawned multiple methods of correction. For the first 40 years of clinical management of this lesion, all methods of correction were surgical. The situation has become more complex with the introduction over the past decade of balloon dilation as a form of management. In search of the ideal form of therapy, numerous comparisons of these various therapeutic methods have been attempted. If we momentarily set aside the distinct possibility that a single ideal form of therapy for all forms of coarctation may well be a misguided concept, we still find that such comparisons have little meaning. The available data in published reports make it literally impossible to objectively compare and contrast these various methods of therapy. There are a number of reasons for this. One of the most important, aside from the intrinsic complexity of the lesion, is that individual studies span widely different time periods (including the preprostaglandin era) and often focus on a particular age group or a particular morphologic subset. To further confuse this situation, outcome assessment after coarctation repair is a quagmire. A "rest" gradient >20 mmHg is the most common measure for determining residual or recurrent coarctation after repair, although there is certainly no consensus on this point. Even if this criterion were universally accepted, the situation would be further confounded by the fact that the gradient may have been determined by any one of several methods, including simultaneous upper and lower body extremity blood pressure measurements using the sphygmomanometer, echocardiographic-Doppler ultrasound flow velocity changes or cardiac catheterization direct pressure measurements. The problem of collateral development also remains after repair and can reduce the validity of the gradient in the same manner as described before for native coarctation. The imprecision involved in defining both the severity of native coarctation and the degree of residual or recurrent lesions creates self-evident problems with outcome assessment and comparison of different repair techniques.

Present study. In the current issue of the Journal, Rao et al. (1) present the results of a 5- to 9-year follow-up period in a series of 67 patients who underwent balloon dilation of native aortic coarctation. The physiologic spectrum and age of these patients run the full gamut. The authors present outcome data

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that, in some cases, appear promising (i.e., results in older children and young adults). However, in other cases, the outcome data raise major concerns, for example, an 83% incidence of "recoarctation" in neonates in association with a 21% incidence of detectable femoral artery injury. A major focus of the study is the development of a comparison between these balloon dilation results and a pooled experience of surgical coarctation repair derived from published reports. The pooled surgical reports span a period of 15 years with respect to their publication dates, and the cases themselves span an even longer time period.

Perspective. It should be pointed out that throughout the history of surgical coarctation repair, discussions focusing on the ideal surgical management of coarctation have been ongoing, and the subject remains controversial to this day. Resection with end to end repair, extended resection, modifications of extended resection, subclavian flap angioplasty and even patch aortoplasty have all had their ardent proponents, yet there remains no consensus of opinion. Amid all this confusion, however, one observation seems to be clear—there are distinct trends toward improved outcome (less mortality and recurrence) in surgical series reported more recently. If there is no consensus regarding the efficacy of various surgical techniques, it would seem to add to the confusion to lump all of these disparate surgical series together to generate a global "surgical" outcome. Furthermore, to then use these outcome statistics as a basis for comparison with another form of management, developed in a more recent era, such as balloon dilation, becomes a meaningless exercise.

Needless to say, to enter into a further statistical argument would, in my opinion, be pointless. What is needed are meaningful data on the various techniques of repair of aortic coarctation using uniform and carefully crafted outcome criteria. This of course means prospective design, randomization and large numbers of subjects accumulated over a relatively short period of time.

I suspect that Rao et al. truly believe in the efficacy of balloon dilation of aortic coarctation; however, I also suspect, at the risk of being presumptuous, that they appreciate the inherent problems in the generation of their "surgical outcome" statistics, as well as the subsequent comparison of these values with the results of their balloon dilation series. If their purpose is to catalyze the development of a meaningful study of the various techniques of aortic coarctation repair, then their strategy may well be effective.

As a final note, I believe that the subject at hand is an excellent example of our collective tendency to derive comfort and security from the generation of statistics even if the application of these statistics is sometimes erroneous. We must recognize when dealing with uncontrolled, complex biologic systems; "filtered" or derived historical data; and poorly defined study and outcome variables that the application of statistical methods will render results meaningless or even misleading.

In the spirit of this message, I close with the following observation. It is clear that the primary caretaker of the patient with congenital heart disease is the pediatric cardiologist. As a result, the cardiologist has the greatest influence in deciding the form of therapy that his or her patient will receive. Catheter intervention techniques for numerous forms of congenital heart disease have been in existence for well over a decade and have been applied to a spectrum of lesions. The pediatric cardiology community has thoroughly embraced the catheter management of certain lesions, such as pulmonary stenosis. However, this same community as a whole has not embraced balloon dilation of native aortic coarctation. The collective voice of the primary caretakers has spoken. The vast majority of patients with native coarctation continue to be referred for surgical correction as the procedure of choice. This choice is based on clinical judgment, with little regard for the available, poorly derived statistics. It is unlikely that the consensus of opinion of this group of caretakers will be influenced to change on the basis of the current narrow focus of both medical and surgical published reports regarding this complex issue.

Reference

1. Rao PS, Galal O, Smith PA, Wilson AD. Five- to nine-year follow-up results of balloon angioplasty of native aortic coarctation in infants and children. J Am Coll Cardiol 1996;27:462–70.